

DIAGNOSTIC VALUE OF CT SCAN IN ORBITAL DISEASES WITH HISTOPATHOLOGICAL CORRELATION

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CERTIFICATE

Certified that this dissertation **entitled “Diagnostic Value Of CT Scan In Orbital Diseases With Histopathological Correlation”** submitted to The Tamil Nadu Dr.M.G.R Medical university, march 2010 is the bonafide work done by Dr.M.KOKILAM, under our supervision and guidance in the Orbit and Oculoplasty Services of Aravind Eye Hospital and Post Graduate Institute of Ophthalmology, Madurai, during her residency period from May 2007 to April 2010

Dr.USHA KIM

Chief, Orbit & Oculoplasty Services
Aravind Eye Hospital
Madurai

DR.M.SRINIVASAN

Director
Aravind Eye Hospital
Madurai

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ANNEXURE

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PROFORMA

MASTER CHART

INTRODUCTION

The technique of CT scanning was originated by Sir Godfrey Hounsfield in England and led to his being awarded the noble prize for medicine in 1979.

PHYSICAL PROPERTIES

When an x ray is transmitted through a substance, the beam is attenuated as a function of both atomic number of the element and the concentration of substances forming the structure. In reality it is the effective electron density that causes attenuation. Increasing the energy of the x ray leads to decrease in attenuation. Conventional x rays are taken with the film alongside the patient and perpendicular to the beam. Most of the rays pass perpendicular to the film; some are scattered in other directions. Because of the thickness of the structures being filmed and the superimposition of these same structures, unwanted shadows are seen in addition to the desired image.

Tomography was invented to eliminate these undesired shadows and to concentrate on the object of interest. In this the x ray source and the film move relative to the patient during exposure. The point at which this source- film plane is pivoted is the object of interest. The desired structure remains motionless, and there is no relative movement between the film and the x ray source.

WINDOWS

The attenuation coefficient was given an arbitrary value by Hounsfield: water was set at 0 and air at -500. On the original scale the dense bone had a typical value of +500. Subsequently, these values were doubled, creating Hounsfield units, abbreviated H, ranging from air at -1000 to bone at +1000. The importance is in providing a numeric matrix from which the computer is able to yield a picture used for diagnostic purposes.

The window level is the central point of the window. In orbital scanning, both soft tissue and bone windows may be required. A central soft tissue window level is usually near 0 to 40 H, with the width of 200 to 400 H. bone windows may have a central level between 40 and 300H, with a width of 2400 to 3200H. This wide window width is necessary because of the variable density of bone.

DEVELOPMENT – CT TECHNOLOGY

The accelerated development of the technology of computed tomographic scanning has led to new generations of advanced scanners.

1. Decreased time required for examination is most important in children and uncooperative patients. Instead of a single beam, a fan beam could be emitted and received by a number of detectors.

2. Increased resolution is obtained by decreasing voxel size through more rapid switching of the detectors. This allows for increased detail at no increased radiation risk.
3. Larger gantry apertures make possible direct coronal scans and certain nonmidline sagittal scans.
4. Thinner sections allow for increased detail.

AXIAL PLANE IMAGING

It is usually related either to orbitomeatal line,OML or Reid's anatomic base lineRBL.OML is a straight line from the lateral canthus to the centre of the external auditory meatus.RBL is a line between the inferior orbital rim and the upper margin of the external auditory meatus. An important angle is the plane of the optic canal, which is -10 degree to RBL and -20 degree to OML. Orbits are usually scanned parallel to RBL or 10 degree negative to OML to achieve an axial plane. For intracranial structures, angulation between 0 degree and +25 degree to OML is useful.

CORONAL PLANE IMAGING

Direct coronal scans require placing the patient in a prone position, with the head rested on the chin, or in a supine position, with the head extended back on its vertex. Coronal scans are taken at 90 degree to RBL. Dental fillings may prevent direct coronal scans; the plane may need to be changed to avoid inducing artifact. Direct coronal images are better than axial images in

assessing the inferior and superior rectus muscles, the optic nerve, optic chiasma, sellar and other parasellar structures.

CONTRAST ENHANCEMENT

Injection of intravenous contrast media is an adjunct to CT scanning. These dyes do not cross the blood brain barrier. Contrast is crucial in the evaluation of chiasmal and parachiasmal lesions. The major complications following use of contrast media are allergic reactions and can be life threatening.

ANATOMY OF ORBIT

The two bony orbits are quadrangular truncated pyramids situated between the anterior cranial fossa above and the maxillary sinuses below. Each orbit is formed by seven bones –

1. Frontal
2. Ethmoid
3. Lacrimal
4. Palatine
5. Maxilla
6. Zygomatic
7. Sphenoid.

The depth of the orbit is 42mm along the medial wall and 50mm along the lateral wall. The base of the orbit is 40 mm in width and 35 mm in height. The volume is 29 ml. The ratio between volume of the orbit and of the eyeball is 4.5 : 1.

WALLS OF ORBIT

MEDIAL WALL

It is formed by the frontal process of maxilla, lacrimal bone, orbital plate of ethmoidal bone and the body of sphenoid.

Relations

- Medial to the medial wall lie anterior ethmoidal air sinuses, middle meatus of nose, middle and posterior ethmoidal sinuses and sphenoidal air sinuses.
- The orbital surface of medial wall is related to superior oblique muscle and medial rectus muscle. In between the two muscles lie anterior ethmoidal nerve, posterior ethmoidal nerve, infra trochlear nerve, terminal branch of ophthalmic artery.

Clinical applications

It is the thinnest wall of the orbit. This accounts for ethmoiditis being the commonest cause of orbital cellulitis, especially in children. The medial wall is frequently eroded by inflammatory lesions, cysts and neoplasms.

INFERIOR WALL

It is formed by three bones: the orbital surface of the maxillary bone medially, the orbital surface of the zygomatic bone laterally and the palatine bone posteriorly. The posterior part of the floor of the orbit is separated from the lateral wall by the inferior orbital fissure. This fissure is continuous anteriorly with the inferior orbital groove.

Relations

- Below it is related to maxillary air sinus.

- Above it is related to inferior rectus muscle, inferior oblique muscle and nerve to inferior oblique.

Clinical applications

The orbital floor being quite thin is commonly involved in 'blow- out fractures' and is easily invaded by tumors of the maxillary antrum.

LATERAL WALL

It is formed anteriorly by the zygomatic bone and posteriorly by the greater wing of the sphenoid bone. More anteriorly the wall is marked by the zygomatic groove and foramina. On the anterior part of the wall is a projection, the lateral orbital tubercle of whitnall. It gives attachment to the check ligament of the lateral rectus muscle and to the suspensory ligament of the eyeball.

Relations

- It separates the orbit from temporal fossa and from middle cranial fossa.
- Medially it is related to lateral rectus, lacrimal nerve and vessels and zygomatic nerve.

Clinical applications

The anterior half of globe is not covered by bone on lateral side. Hence, palpation of retrobulbar tumors is easier from the lateral side. It is the strongest portion of the orbit and needs to be sawed open in lateral orbitotomy. The zygomatico- sphenoid suture is an important landmark during surgery.

ROOF

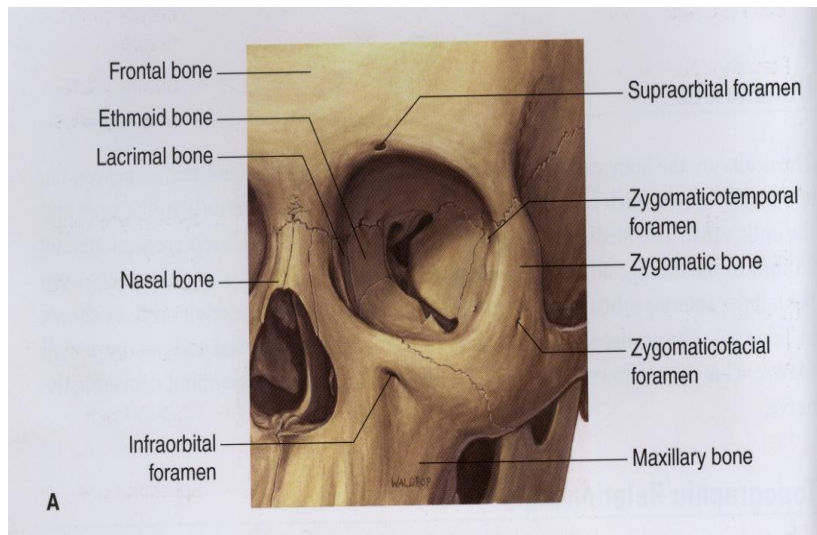
It is formed by the orbital plate of the frontal bone, the lesser wing of sphenoid. The anterolateral part of the roof has a depression called the fossa for the lacrimal gland. The troclear fossa is situated at the junction of roof and the medial wall.

Relations

- Above, it is related to frontal lobe cerebrum and meninges.
- Below, it is related to frontal nerve, levator palpebrae superioris, superior rectus, superior oblique, trochlear nerve and lacrimal gland.

Clinical applications

As the roof is perforated neither by major nerves nor by blood vessels, it can be easily nibbled away in transfrontal orbitotomy.



SURGICAL SPACES IN ORBIT

The orbit is divisible into a number of spaces. Knowledge of the main compartments of the orbit and their boundaries helps in choosing the most direct approach to the tumor.

SUBPERIOSTEAL SPACE

This is a potential space between orbital bones and the periorbital. Dermoid cyst, epidermoid cyst, mucocele, subperiosteal abscess, myeloma, osteomatous tumor, hematoma and fibrous dysplasia are commonly seen in this space.

PERIPHERAL ORBITAL SPACE (ANTERIOR SPACE)

This space is bounded peripherally by periorbital, internally by the four extra ocular muscles, anteriorly by the septum orbitale. Posteriorly, it merges with the central space. Tumors present in this space produce eccentric proptosis

and can usually be palpated. Common tumors present in this space are malignant lymphoma, capillary hemangioma, neoplasms of the lacrimal gland and pseudotumors. Tumors residing in this space are explored mostly by anterior orbitotomy.

CENTRAL SPACE

It is also called muscular cone or posterior or retrobulbar space. Contents of this space include optic nerve and its meninges, superior and inferior divisions of oculomotor nerve, abducent nerve, nasociliary nerve, ciliary ganglion, ophthalmic artery, superior ophthalmic vein and central orbital fat. Many of the circumscribed orbital tumors such as cavernous hemangioma, solitary neurofibroma, meningioma, optic nerve glioma occur in this space and usually produce axial proptosis. Such tumors are often removed through a lateral orbitotomy.

SUB- TENON’S SPACE

It is a potential space around the eyeball between the sclera and tenon’s space. Pus collected in this space is drained by incision of tenon’s capsule through the conjunctiva.

APICAL SPACE

It is bounded peripherally by periorbita; anteriorly becoming continuous with the anterior and central spaces.

NORMAL CT ANATOMY

Axial and coronal views are complementary for showing bony and soft tissue anatomy. The axial view is superior for demonstrating the lateral and medial bony margins, the superior orbital fissure, and the optic canal. Coronal views are best for assessing the floor and roof. The lacrimal sac and nasolacrimal duct as well as the inferior orbital fissure and infraorbital canal are equally seen on axial or coronal images.

The optic nerve has a serpiginous course with minimal inferior and lateral bowing in its midportion. Because of this, thin slices may not show the entire course of the nerve on any one axial slice. The nerve can be well defined throughout its course, except within optic canal. The dural sheath along the optic nerve is well defined with intravenous contrast. On coronal views immediately posterior to the globe, a small central density within the nerve represents the central retinal artery and vein.

The extraocular muscles generally have a course parallel to the adjacent orbital wall. consequently only the medial and lateral recti are seen in their entirety on an axial view. The superior and inferior recti are only partially visualized on any axial slice and on coronal views are seen as cross sectionally to lie in a slightly oblique plane. The levator palpebrae superiors merges with

the superior rectus and is only identified separately on anterior coronal images where it diverges from superior rectus. The superior oblique is best seen on coronal views lying superior and slightly medial to medial rectus. The least well defined muscle is the inferior oblique, with only its insertion well seen on axial views.

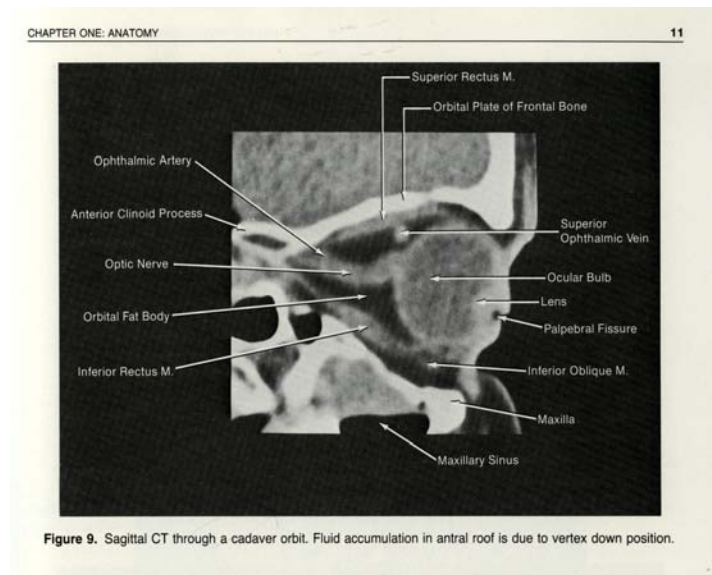
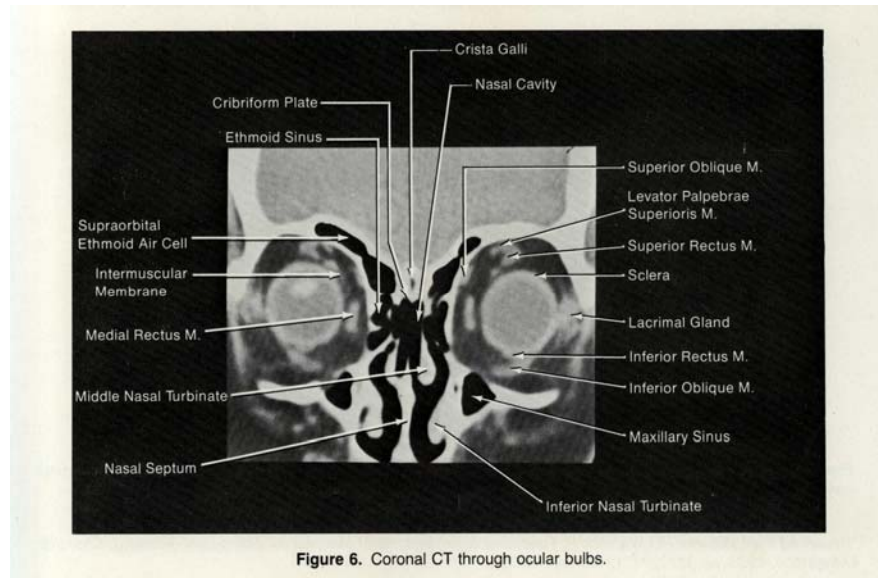
The lacrimal gland is readily identified in lacrimal fossa on both coronal and axial views. The superior portion of nasolacrimal duct can be readily identified on axial and coronal views.

The vascular structures in the orbit can be seen without intravenous contrast, but are highlighted with contrast. The ophthalmic artery is seen in the apex of orbit. Several of its branches, including the anterior and posterior ciliary branches can usually be identified. The superior ophthalmic vein is routinely identified in axial and coronal views as it course near trochlea to pass through the muscle cone inferior to the superior rectus and superior to optic nerve to exit the orbit through the superior orbital fissure.

A line joining the lateral orbital margins in axial plane will normally intersect the globe near its midportion, with atleast one third of the globe posterior to this line. The sclera, choroid and retina form a well defined band that enhances with intravenous contrast. The lens is normally high density on CT.

Nerves can occasionally be identified in orbit.

The cavernous sinus is particularly well seen with contrast. The third, fourth, first division of fifth nerve and sixth nerve appear as round, low density structures on coronal views through the uniformly enhancing cavernous sinus.



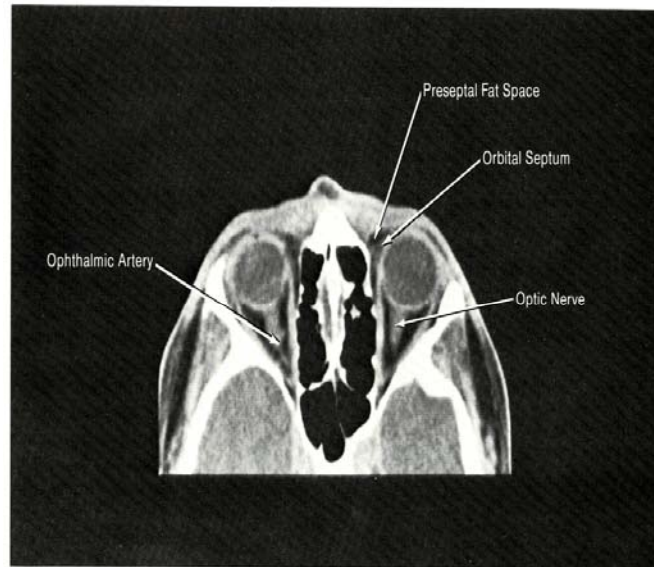


Figure 10. Axial CT defining preseptal space.

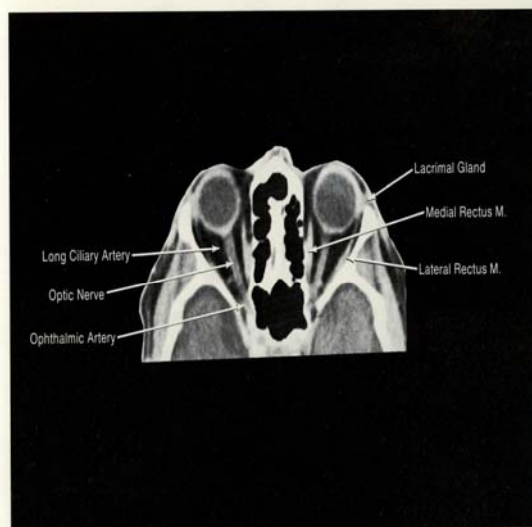


Figure 12. Detail of the optic nerve. The central linear lucency represents the optic nerve surrounded by a denser optic nerve sheath.

ANATOMIC PATTERNS OF ORBITAL DISEASES

The effect of any disease in the orbit is not only governed by the primary nature of the process but also by the anatomic pattern of involvement.

The patterns of anatomic involvement can be divided into anterior, diffuse, apical, myopathic, ocular, intraconal, optic nerve, periorbital and lacrimal drainage system.

ANTERIOR

The anterior inflammations may be characterized by pain, diplopia, chemosis, lid swelling, injection, uveitis, papillitis, optic neuropathy, and even exudative retinal detachment. Characteristically, on CT investigation a contrast enhancing anterior orbital infiltration intimately related to the globe produces scleral and choroidal thickening.

DIFFUSE

It is similar in presentation to anterior, but is more severe. It is frequently associated with optic neuropathy, motor and sensory deficits.

APICAL

It produces less proptosis, pain or visible inflammation, but is associated with early development of optic neuropathy, motor or sensory symptoms. In addition, when the process affects the superior orbital fissure at the apex, vascular congestion due to the obstruction of the superior or inferior ophthalmic

veins may be a clinical feature. Thus the patient reports pain, limitation of movement or visual deficit.

MYOPATHIC

It is characterized by pain with eye movement, localized injection of globe over the insertion of affected muscle and limited ocular motility. On CT scan, the contrast enhancing irregular infiltrate involves one or more extraocular muscles with relatively diffuse enlargement extending upto the globe and usually including the tendon.

OCULAR

Ocular inflammation can extend to involve surrounding orbital structures.

PERIORBITAL

Diseases originating from the sinuses, face and intracranial cavity may extend into the orbit by contiguity or as a result of damage to neurosensory or vascular structures shared with the orbit.

INTRACONAL

Lesions within the muscle cone produce axial displacement and functional deficit of the eye, optic nerve, muscles and ciliary ganglion.

OPTIC NERVE

The effect of the disease is governed by whether the process is intrinsic to the nerve or affects the nerve from the sheath.

LACRIMAL

Acute idiopathic inflammation presents with localized pain, tenderness, injection of the temporal lid and fornix with palpable lacrimal gland, S shaped deformity of lid.

COMMON ORBITAL DISEASES

OPTIC NERVE GLIOMA

Gliomas are of glial origin. The majority are isolated lesions, but a significant proportion arise within the context of neurofibromatosis. Bilaterality is pathognomonic for Von Recklinghausen 's disease. There is female predominance, with 75% occurring in the first decade. The clinical character can be best understood by dividing into orbital, orbito cranial, chiasmal and diffuse type.

Orbital type

They arise within the orbit where they are characterized by proptosis (frequently non axial) and visual loss.

Orbitocranial type

They fall into three sub groups: anterior to chiasm, chiasmal involvement, para chiasmal involvement.

Diffuse type

They occur in patients with von recklinghausen's disease.

CT

Orbital type presents as enlarged fusiform optic nerve with smooth intact dural margins. Anterior kinks adjacent to the globe and low density cystic areas

are frequent and characteristic, reflecting pliability of the nerve and focal areas of degeneration within the tumor.



Orbitocranial type – high resolution axial and coronal CT scans with thin cuts are mandatory to evaluate para chiasmal region.

Diffuse type – multifocal or diffuse enlargement of the optic nerve is seen.

HISTOPATHOLOGY

They are well differentiated pilocytic astrocytoma. Two growth patterns have been described. The majority arise intrinsically, expanding the individual fascicles and the overall dimension of the nerve. The second pattern is extra neural extension in the arachnoid space and is believed to be more common in neurofibromatosis. There is hyperplasia of the surrounding arachnoidal cells, which may lead to an incorrect diagnosis of meningioma. However, gliomas

characteristically do not invade the dura. Vascular proliferation and atypia are common and do not indicate malignancy.

MANAGEMENT

The general principle of management is essentially conservative. If it is limited to the orbit observation is recommended unless there is disfiguring proptosis or significant progression. When the tumor is present anterior to the chiasm but not involving the chiasm requires more prompt surgical action because once the chiasm is involved excision is impossible. This subgroup can be managed by surgical excision of the involved nerve up to the chiasm with confirmation of clear margins. Patients with chiasmal involvement represent the most distressing group, because there is no reliable and definitive therapy. Intervention can even ablate residual vision. Non-surgical treatment such as radiotherapy and chemotherapy remain controversial. In diffuse type, several reports have indicated a good visual prognosis even without intervention.

MENINGIOMA

They frequently lead to visual loss; those confined to and arising from the optic nerve sheath cause unilateral deterioration, whereas those arising intracranially often affect vision bilaterally.

Intracranial meningioma

The major sites affecting the orbital and visual structures are the sphenoid ridge, suprasellar area and olfactory groove. The medial tumors cause

cranial nerve palsies, visual deficits and venous obstruction. Remote tumors exert their effect by virtue of raised intracranial pressure or mass effect.

Optic canal meningioma

It typically presents with visual loss due to early compression of the optic nerve.

Optic nerve meningioma

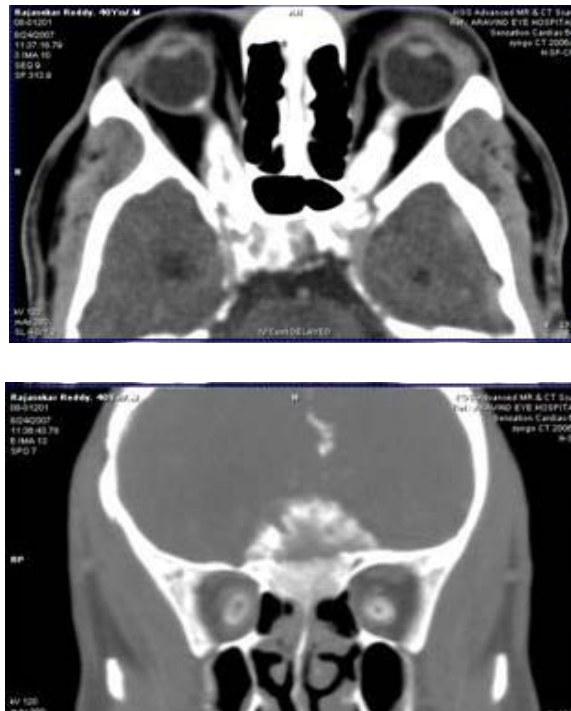
The majority occur between third and sixth decades with about two thirds of cases affecting females. The clinical features are slowly progressing compressive optic neuropathy, transient obscuration of vision, mild proptosis.

CT

Intracranial – lesion is well defined, homogenous and characteristically of increased density with uniform enhancement post contrast infusion.

Optic canal meningioma- even with the use of thin sections it may be very difficult to identify small tumors of optic canal.

Optic nerve meningioma generally demonstrates one of the three radiologic patterns: diffuse thickening, fusiform swelling anterior or posterior globular enlargement. Often the border is nodular. Central lucent areas after contrast infusion are characteristic and may identify the residual optic nerve. There is a pathognomonic subgroup characterized by calcification and railroad tracking. Intracranial extension may be noted as small tumors in the region of the anterior clinoids.



HISTOPATHOLOGY

The meningotheelial cap cells of the arachnoid villi are considered stem cells of meningioma. Infiltration of bone can cause hyperostosis, which may make meningiomas difficult to distinguish clinically from primary bone tumors. The fundamental cell is usually round or polygonal, but may be more spindle shaped. Two thirds of tumors are meningotheliomatous, consisting of whorls of spindle cells that frequently surround a central psammoma body. Dense, randomly woven bundles of spindle shaped meningotheelial cells and fibroblasts constitute the fibrous pattern. Two aggressive patterns exist: the angioblastic and sarcomatous.

MANAGEMENT

Patients can be observed as long as there is little progression or evidence of intracranial involvement. When confined to anterior two thirds of the optic nerve in orbit, the optic nerve and meningioma may be excised through lateral orbitotomy. If there is apical or intracranial involvement, a combined neurosurgical ophthalmic panoramic orbitotomy may be needed.

SCHWANNOMA

They are well defined, encapsulated, slowly progressive tumors that arise from peripheral nerves. They are usually solitary, occur between the ages of 20 and 50 years.

CLINICAL FEATURES

Intraconal

- * proptosis
- * lid swelling
- * diplopia in extremes of gaze.
- * central scotoma can occur in apical tumors.

Extraconal can occur anywhere in and around orbit, including sinuses, lacrimal sac, etc

CT

It demonstrates well defined, smooth, rounded contours, with variation in density based on cyst formation, degeneration and lipid deposition in some tumors. With contrast injection, mild enhancement may be noted.



HISTOPATHOLOGY

They consists of proliferations of schwann cells within a perineural capsule that displace (or) compress the nerve of origin. The characteristic pathology is an admixture of tightly ordered schwann cells(Antoni A area) and a loosely arranged component(Antoni B area) within the capsule. They contain less acid mucopolysaccharaide than neurofibromas. Because they are slow growing and late in presentation, features of degeneration may dominate in large masses. These include cyst formation, hemorrhage, calcification, hyalinization, infiltration with siderophages and lipid laden schwann cells.

MANAGEMENT

Large or symptomatic tumors can be managed by surgery, the approach being governed by location. At surgery, they are characteristically yellow – tan, solid, encapsulated and have typical varicose, violaceous tumor vessels on the surface. Removal may be total or subtotal, in piecemeal fashion or by evacuation of tumor within its capsule. Evacuation of these tumors may be wise if located in critical sites and can be aided by using ultrasonic fragmentation suction devices.

NEUROFIBROMA

Plexiform neurofibroma

They are the most common complex peripheral nerve tumors of the orbit.

Clinical features

The overlying skin may be thickened. The tortuous, ropey, tangled nerves produce a characteristic palpable bag of worms. The soft tissues of lid, periorbita and face are thickened, hypertrophied or even pendulous producing varying degree of proptosis and facial disfigurement. The patients may have uveal neurofibroma(lisch nodule) or choroidal lesions.

Solitary neurofibroma

These lesions tend to be seen in middle- aged persons.

Clinical features

It manifests as solitary masses with preponderance of occurrence in the upper quadrant. Clinically, they are solid, isolated, circumscribed masses. As they chiefly affect sensory nerve, anesthesia, paresthesia, hypesthesia can occur.

Diffuse type

It is a rare dermal form characterized by infiltration and envelopment of normal structures.

CT

In Plexiform neurofibroma – contrast enhancing irregular soft tissue infiltration, extra ocular muscles involvement may be seen. Retrobulbar fat may show increased density. Bony changes consists of enlargement of the orbit, widening of superior and inferior orbital fissures, hypoplasia of the ethmoid and maxillary sinus, defects in greater wing of sphenoid and enlargement of middle cranial fossa.

Solitary type - it appears as well circumscribed , usually homogenous mass.

Diffuse- there is hyper density of fat, muscles and soft tissues.



HISTOPATHOLOGY

They may not be well encapsulated and contain loosely arranged interlacing bundles of spindle cells and collagen fibrils within mucoid matrix.

MANAGEMENT

Plexiform type- cosmetic surgery, consisting of repeated debulking or orbital bony enlargement.

Solitary and diffuse type- surgical excision.

CAVERNOUS HEMANGIOMA

They are benign noninfiltrative lesions.

CLINICAL FEATURES

These tumors are typically intraconal.

- * proptosis

- * posterior pole indentation

* choroidal striae

CT

It shows very well defined oval or rounded intraconal mass with smooth margins, which enhances with contrast. Occasionally, a small portion may extend into the extra conal compartment, but the greatest bulk is almost always intraconal. The enhancement pattern may be homogenous or inhomogenous with approximately equal frequency. The nerve is typically displaced rather than surrounded by the tumor.



HISTOPATHOLOGY

It reveals a fine capsule that surrounds a tumor consisting of large endothelially lined channels with abundant, loosely distributed smooth muscle in the vascular walls and stroma.

MANAGEMENT

Surgical excision is the treatment of choice. When exposed, the tumor is plump, nodular, plum coloured mass with vascular channels on its surface. It

can be removed by blunt dissection of the surface to free it of adjacent orbital structures. There is frequently an apical vascular twig, which is best left to the final stage of the procedure because rupture earlier can cause more bleeding and obscure the field.

PLEOMORPHIC ADENOMA

They usually occur in the second decade to fifth decades, with the peak incidence in the fourth.

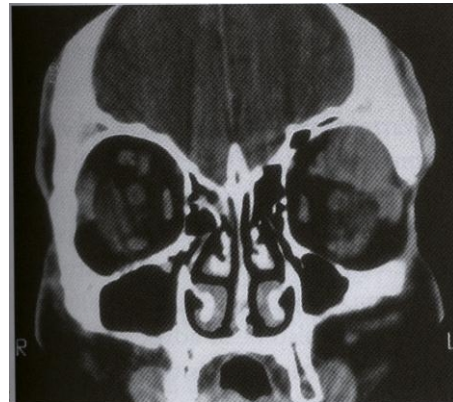
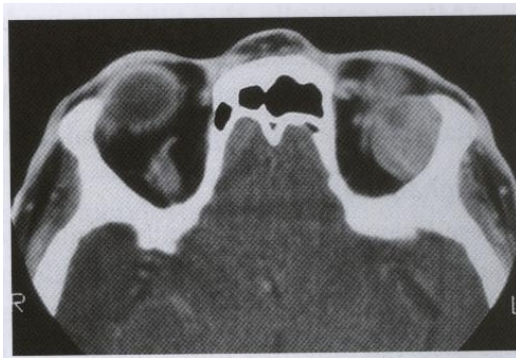
CLINICAL FEATURES

They may present as progressive painless downward and inward displacement of the globe. Large tumors may be associated with blurring of vision, diplopia, retinal and choroidal striae.

CT

Well circumscribed hyperdense mass in the superolateral quadrant of orbit is present.

Enlargement or expansion of lacrimal fossa may be present due to pressure erosion.



HISTOPATHOLOGY

On gross pathological examination they appear as grayish- white, bosselated, solitary mass that are well circumscribed by a pseudocapsule. The pseudocapsule is a deceptive component of the lesion, and the histology shows excrescences invading this condensation. It is this feature that has led to a high incidence of recurrence when a margin of a normal tissue is not excised. There are two morphological components-one is composed of cells resembling ductal epithelium, whereas the other consists of stellate, spindle cells streaming in loosely arranged stroma. The stromal component may be myxoid, hyalinized, pseudocartilaginous or calcified.

MANAGEMENT

Extirpative biopsy by a modified lateral orbitotomy can be done. The prognosis is excellent. The recurrence following incomplete excision may take a long time and may be associated with malignant transformation.

CARCINOMA AND PLEOMORPHIC ADENOMA MALIGNANT MIXED TUMOR)

There are three clinical circumstances in which malignant mixed tumors arise.

- Sudden expansion of an indolent long standing lacrimal mass
- Pain, bony infiltration and rapid growth.
- Sudden recurrence of a previously excised mixed tumor of lacrimal gland.

MANAGEMENT

When these tumors arise as a component of a benign mixed tumor they are usually approached by a lateral orbitotomy and en bloc excision. If malignant transformation is discovered locally within the mass, a wider excision of adjacent tissues must be carried out. The role of radiotherapy has not been clearly established in the treatment of this tumor.

ADENOID CYSTIC CARCINOMA

It is the most common epithelial carcinoma of the lacrimal gland. It occurs in either sex with a peak in the fourth decade.

CLINICAL FEATURES

Eccentric proptosis, increased tearing, double vision, pain, firm mass in the superotemporal quadrant.

CT

Tumor mass may be less circumscribed. There may be areas of pressure erosion, bony destruction, or calcification in the lacrimal fossa.



HISTOPATHOLOGY

The most distressing aspect of the fundamental pathology of this carcinoma is its tendency to perineural invasion. Grossly, it is grayish- white with a firm nodular surface. Microscopically, the cell population consists of densely packed hyperchromatic small cells with scant cytoplasm. The cystic spaces vary considerably in size and number and may be so numerous to give swiss cheese pattern. On the other hand, the tumor may be dominantly cellular, forming solid cords. Five histological patterns have been described: swiss cheese pattern, sclerosing, basaloid, comedocarcinomatous and tubular.

MANAGEMENT

When the tumor appears to be confined to the orbital tissues the recommended therapy is en bloc excision by a multidisciplinary team. Radical radiotherapy is often recommended as an adjunctive measure, but is not of proven efficacy.

LYMPHOMA

It forms the largest group of lymphoproliferative disorders of orbit. The onset is characteristically in the sixth or seventh decades of life.

CLINICAL FEATURES

They usually occur in the anterior orbit and may be associated with pink, fleshy sub conjunctival tumefaction, which tends to mold to the shape of the orbit. When there is no visible subconjunctival component, these lesions tend to be palpably nodular with relatively well defined margins.

CT

These lesions are usually fairly well defined and tend to encompass adjacent ocular and orbital structures. They almost and always have an extra conal component; an associated intra conal component may be seen, but rarely occurs alone. Lacrimal gland involvement is common and may be the only site of tumefaction. They almost universally involve the orbital soft tissues, but rarely can be seen within an extra ocular muscle. They are isodense to the

extraocular muscles both on contrast and non contrast scans. Orbital bony involvement and distortion of globe are rare.



HISTOPATHOLOGY

They are low to intermediate in grade with predominance of low grade tumors. They usually consists of poorly formed follicles with heterogenous cellular composition, including small atypical cells, small lymphocytes, and plasma cells.

MANAGEMENT

Lesions that are localized to the orbit can be treated with orbital radiotherapy or observed until more widespread disease develops. The less well differentiated tumors are treated with systemic chemotherapy or low dose(1500- 2500 rad) radiotherapy or both.

SINONASAL MALIGNANCY

Epithelial malignancies of paranasal sinuses frequently spread to orbit.

By definition, orbital involvement reflects an advanced stage.

CLINICAL FEATURES

- Upward displacement of globe
- Fullness of lower lid
- Infra orbital pain
- Paresthesia
- Distortion of maxilla
- Nasal obstruction
- Epistaxis

CT

Hyperdense lesion with the erosion of adjoining bony walls.



MANAGEMENT

Wide excision

ASPERGILLOSIS

Aspergillus is best known as a source of opportunistic infections.

CLINICAL FEATURES

- Necrotizing angitis due to microscopic foci of fungus in small vessels, leading to thrombosis. Endophthalmitis is a common sequel to this.
- Sclerosing infiltrative mass usually originating from an adjacent sinus. When anterior, it leads to proptosis, but apical infiltrates may cause orbital apex syndrome.
- Rarely focal abscess and fistula formation can occur.

CT

It reveals thick allergic mucus as mottled areas of hyper density. Bone destruction and remodeling are usually present but donot signify actual tissue invasion.



HISTOPATHOLOGY

It reveals thick peanut butter like or green mucus, pathologic study of which reveals numerous eosinophils, eosinophilic degradation products and extra mucosal fungal hyphae.

MANAGEMENT

Local disease is treated with surgical drainage and debridement. Amphotericin B and flucytosine may be used in non neutropenic patients who have invasive aspergillosis.

CYSTICERCOSIS

It is a disseminated infection caused by the larval stage of (cysticercus cellulosae) of the pork tape worm Taenia solium. Humans are the definitive host and pigs ingest eggs passed in human feces.

CLINICAL FEATURES

CNS- headache, seizure, neurological deficit.

OCULAR

RD

Retinal oedema

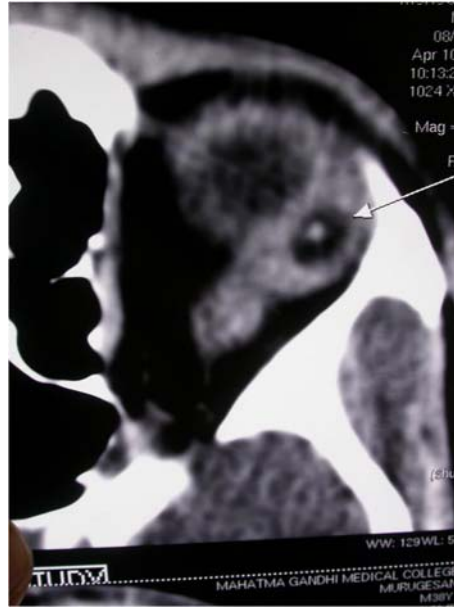
Chorioretinitis

Vitritis

It can occur in periorbital muscles, conjunctiva, sub retinal space and lacrimal gland.

CT

Presence of scolex is a characteristic feature.



MANAGEMENT

Albendazole is the drug of choice. Because anticysticercal agents cause profound inflammatory reaction around dying cysts, the concomitant use of steroids is often recommended.

SPECIMEN HANDLING

ORIENTATION

Globes are oriented according to the location of extra ocular muscles and of the long posterior ciliary artery and nerve, which are located in horizontal meridian. Locating the inferior oblique muscle is very helpful in distinguishing between right and left eye. Once the laterality is determined, the globe may be trans illuminated and dissected.

TRANSILLUMINATION

Eyes are transilluminated with bright light prior to gross dissection. This helps to identify lesions such as intra ocular tumor that blocks the trans illuminated light and casts a shadow. The shadow can be outlined with a marking pencil on the sclera. This outline can be used to guide the gross dissection of the globe so that the center of the section will include the maximum extent of the area of interest.

GROSS DISSECTION

A globe is opened so as to display as much of pathologic change as possible on a single slide, PO section. The meridian, or clock- hour, of the section is determined by the unique features of the case, such as a presence of an intra ocular tumor or a history of trauma or previous surgery. In routine

cases, most eyes are opened in the horizontal meridian, which includes the macula in the same section as the pupil and optic nerve. Globes with a surgical or non surgical wound should be opened so that the wound will be perpendicular to the section, which means opening the globe vertically. Globe with intra ocular tumors are opened in a way that places the center of the tumor as outlined by transillumination in PO section.

FIXATIVES

The most commonly used fixative is 10% neutral buffered formalin. It prevents postmortem enzymatic destruction of tissues. In specific instances, other fixatives such as glutaraldehyde for electron microscopy and ethyl alcohol for cytologic preparations are used. Formalin diffuses rather rapidly through tissue. Because most of the functional tissue is within 2-3 mm of the surface, it is not necessary to open the eye. The adult eye measures about 24mm in diameter, and formalin diffuses at the rate of 1mm/hr; therefore, globes should be fixed at least 12hours prior to processing. It is generally desirable to suspend an eye in formalin in a volume of approximately 10:1 for at least 24 hours prior to processing to ensure adequate fixation.

TISSUE PROCESSING

The infiltration and embedding process removes most of the water from the tissue and replaces water with paraffin. Specimens are routinely processed through increasing concentration of alcohol followed by xylene. Alcohol dehydrated tissue and xylene replaces alcohol prior to paraffin infiltration. The paraffin mechanically stabilizes the tissues, making possible cutting of sections.

TISSUE STAINING

Tissue sections are usually cut at 4-6 micron. The cut section is colorless. Principally hematoxylin and eosin are used to color the tissue for identification. A small amount of resin is placed over the stained section and covered with a thin glass cover slip to preserve it.

REVIEW OF LITERATURE

S.Wende in his study of 520 cases of orbital lesions said that there is considerable improvement and greater accuracy in diagnosis in the field of ophthalmology by using CT. Early tumor visualization is possible without risk or discomfort to the patient.

Hu yanhua's study of 117 cases of orbital tumors revealed that the sensitivity in the diagnosis of orbital tumors by CT was 93.3%. The coincidence of CT diagnosis with HP were 83.3%, 82.2% and 71.4% for dermoid cysts, hemangioma and pseudotumor respectively, but the general coincidence was 67.8%.

In R.P.muller's study of 39 cases of orbital tumors, the clinical use of CT was analysed. CT, clinical and histopathological findings were compared. CT was found to valuable in outlining the tumor itself and its extension to neighboring structures. This will help the surgeon to decide on the optimal approach. In most cases, a preoperative decision was possible as to whether a multidisciplinary team is needed.

Moon and his co- workers conducted a study on 19 histopathologically proven orbital lymphoma and 9 pseudotumor. The differentiation between orbital lymphoma and pseudotumor is difficult clinically and radiologically. The study concluded that different characteristicss of attenuation change on two

phase helical CT and delayed coronal CT can be helpful in differentiating between orbital lymphoma and pseudotumor.

Kelvin studied on metastatic orbital tumor from gastric carcinoma CT showed well defined enhancing intra conal mass. Histopathology confirmed a metastatic adenocarcinoma. Chemotherapy was initiated with good tumor response. He concluded that early biopsy of unusual orbital tumor is critical as orbital metastasis may be the initial manifestation of an asymptomatic primary. Histopathology can aid localization of the primary tumor and allow prompt treatment to be initiated.

Chung huwan back and his co- workers analyzed 18 PET/CT and CT scans in 15 patients with biopsy proven periorbital malignancies. PET/CT had a sensitivity of 100%, while CT had a sensitivity of 57% for nodal metastasis by level- by- level analysis. PET/CT had a specificity of 97%, positive predictive value of 93%, diagnostic accuracy of 98%, while CT values for these parameters were 97%, 89%, 82%, 84% respectively. PET/CT could provide useful information in the management of regional lymph node metastasis in patients with periorbital malignancies.

Anzai.Y conducted a prospective cohort study in a medical center to determine the impact of sinus CT on treatment decisions by otolaryngologist and to explore the factors leading to choice of surgical management for patients suspected of having chronic sinusitis. Treatment decisions were changed in

9/27 patients after sinus CT scans were reviewed. Agreement of treatment decisions among three surgeons was improved after they reviewed sinus CT scans. Study concluded that decision to perform surgery was changed by CT in a substantial number of patients.

Z.A.Sherazi, CR Jayakumar conducted a study to assess the importance of CT in evaluation of retinoblastoma. They reviewed 13 cases of retinoblastoma which presented at hospital university Sains Malaysia, Kelatan, Malaysia from august 1986- June 1991. High resolution CT of orbit was performed in all patients prior to therapy. 69% had unilateral disease and 31% had bilateral retinoblastoma. The interesting features were remarkably high incidence in right eye (89%) as compared to left eye (11%) in unilateral retinoblastoma. CT detected intra ocular calcification in 82% of tumorous eyes. All patients presented at late stages when tumors were of large size. The presence of calcification was not related to the size of tumor. CT detected calcification in suspected retinoblastoma with a high degree of accuracy. CT evidence of intra ocular calcification in children under three years is highly suggestive of retinoblastoma.

Zhonghua Ya Ke and his co-workers investigated histopathologic classification and distribution of orbital diseases. They analysed 3476 orbital diseases examined in 1976-2000 in pathology laboratory. Benign orbital diseases were 81.9%. The ten leading benign orbital diseases were cavernous

hemangioma (515), vascular leiomyoma (364), pseudotumor (347), dermoid (230), schwannoma (183), meningioma(150), benign mixed tumor of lacrimal gland (147), mucocele (141), varix (132), neurofibroma (76). Malignant orbital tumors were 18.10%. The malignant tumors were malignant lacrimal gland epithelial tumor (129), rhabdomyosarcoma (75), non- hodgkin's lymphoma (65), secondary to sinonasal carcinoma (51), metastatic (50), choroma (32), extra ocular extension of retinoblastoma (26) and extra ocular extension of choroid melanoma (23). They concluded that the vascular tumors and malformations are commonly seen in orbital diseases. Primary malignant tumor in orbit is malignant lacrimal gland epithelial tumor.

Mehran Midia and his co- workers examined the accuracy and efficacy of pre -operative imaging by CT and MRI in assessing tumor invasion of orbit. 98 pre- operative CT and 40 pre-operative MRI images from patients with orbital masses were reviewed. Results were corroborated by pathologic and intra- operative assessment. 60.9% of patients were male and 39.1% were female. Sensitivity of CT scan in diagnosis of orbit masses was 89.9% and MRI sensitivity was 95%. Both CT and MRI have high potency in diagnosis of orbit lesions and from the view point of statistics, no significant difference was found between diagnostic accuracy of orbit lesions by CT and MRI ($p>0.005$).

MV Saeed and his co-workers did a retrospective review of all enucleated/ eviscerated histopathology reports over 20 years. Clinical history

was correlated with pathologic findings. Two ten year period (1984-1993) and (1994-2003) were compared to detect changes in incidence of eye removal. The number of eyes removed and histologically analyzed decreased in the period 1994-2003, perhaps because of better diagnostic modality like CT and treatment options available allowing globe preservation.

AIMS AND OBJECTIVE

1. To evaluate diagnostic precision of CT scan in orbital diseases.
2. To establish correlation between CT scan diagnosis and histopathological diagnosis in orbital diseases.

METHODOLOGY

This study was conducted on 100 patients during a period of two years (September 2007-september 2009), who were suspected to have orbital disease. Detailed history was taken and thorough clinical examination was done. CT scan was taken in all patients and all patients underwent appropriate surgical procedure.

CT SCAN - ORBIT

Axial and coronal sections were taken. CT – brain/chest were taken in selected patients in order to achieve and augment the diagnostic accuracy.

SURGICAL PROCEDURE

Most of the patients underwent excision biopsy. Some underwent incision biopsy/ Enucleation/ debulking depending on the diagnosis.

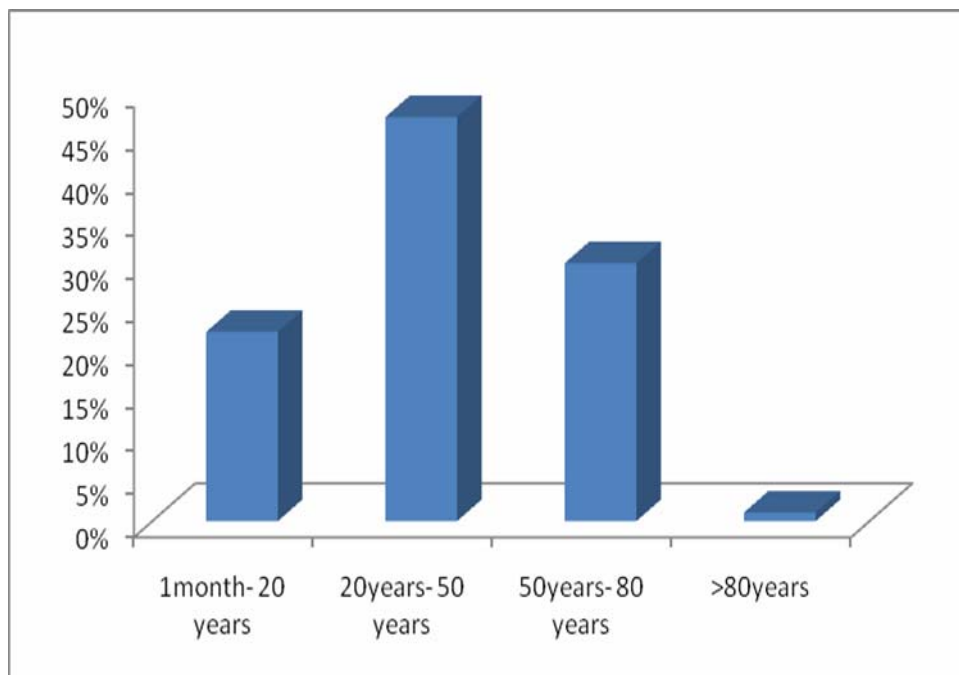
HISTOPATHOLOGY

All the specimens were sent for histopathological examination. Special stains were used whenever needed.

RESULTS

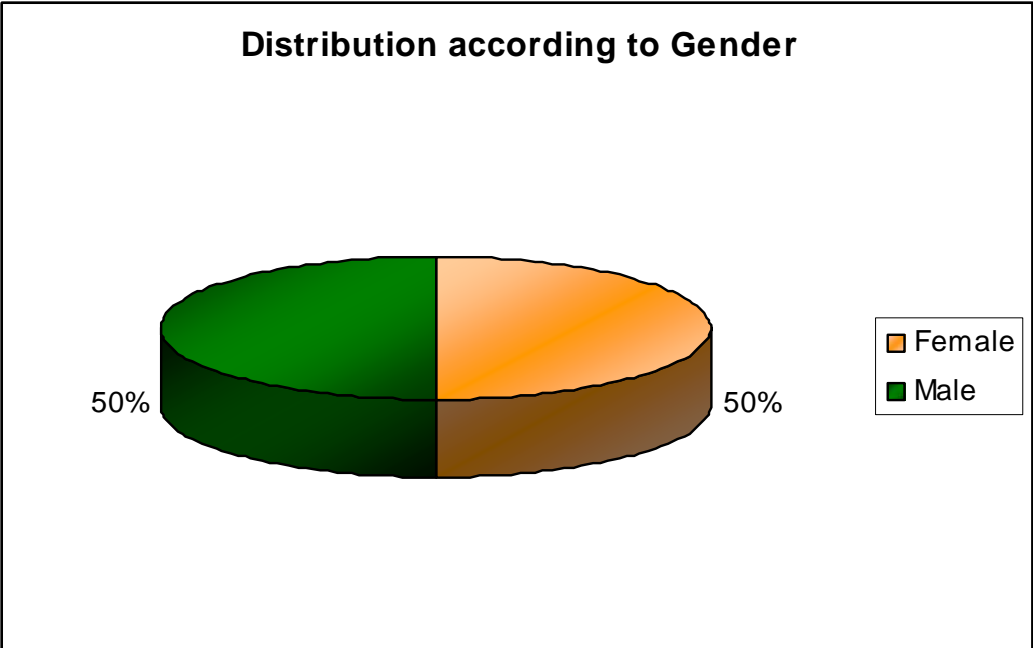
AGE DISTRIBUTION

1month- 20 years	22%
20years- 50 years	47%
50years- 80 years	30%
>80years	1%



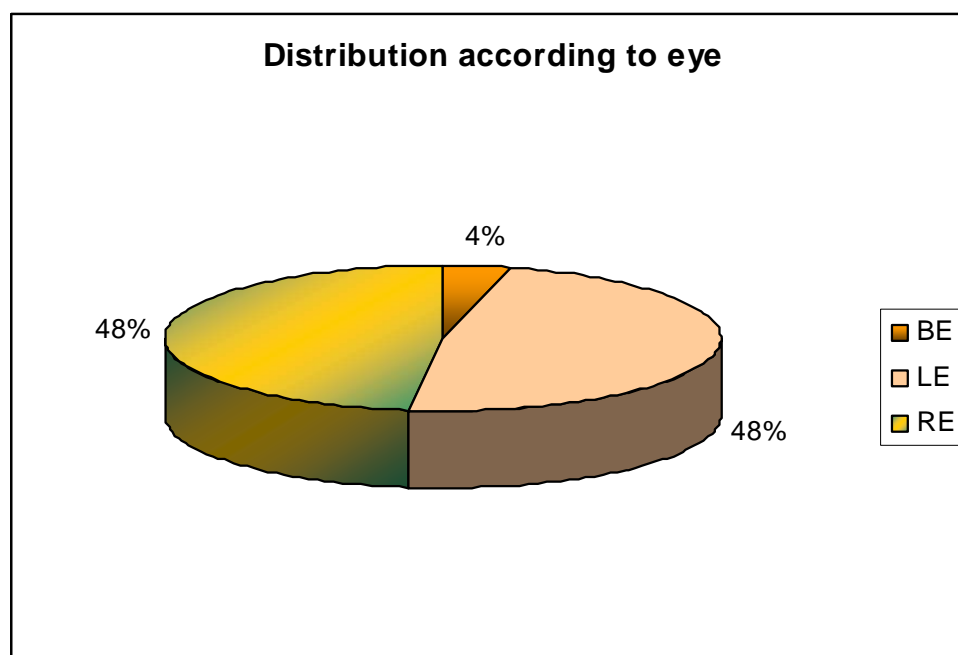
SEX DISTRIBUTION

MALES	50%
FEMALES	50%



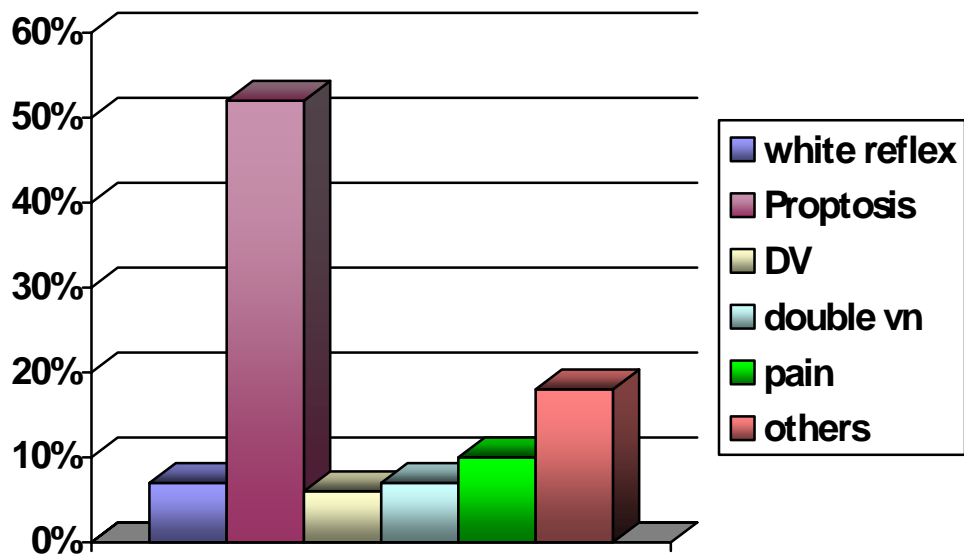
LATERALITY

RIGHT EYE	48%
LEFT EYE	48%
BOTH EYES	4%



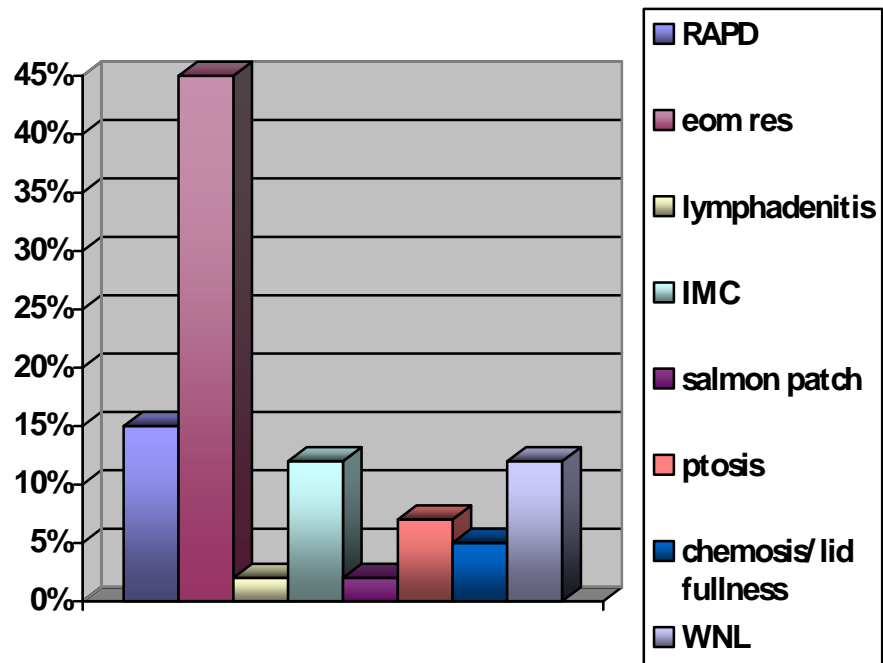
COMPLAINTS

White reflex	7%
Proptosis	52%
Defective vision	6%
Double vision	7%
Pain	10%
Others	18%



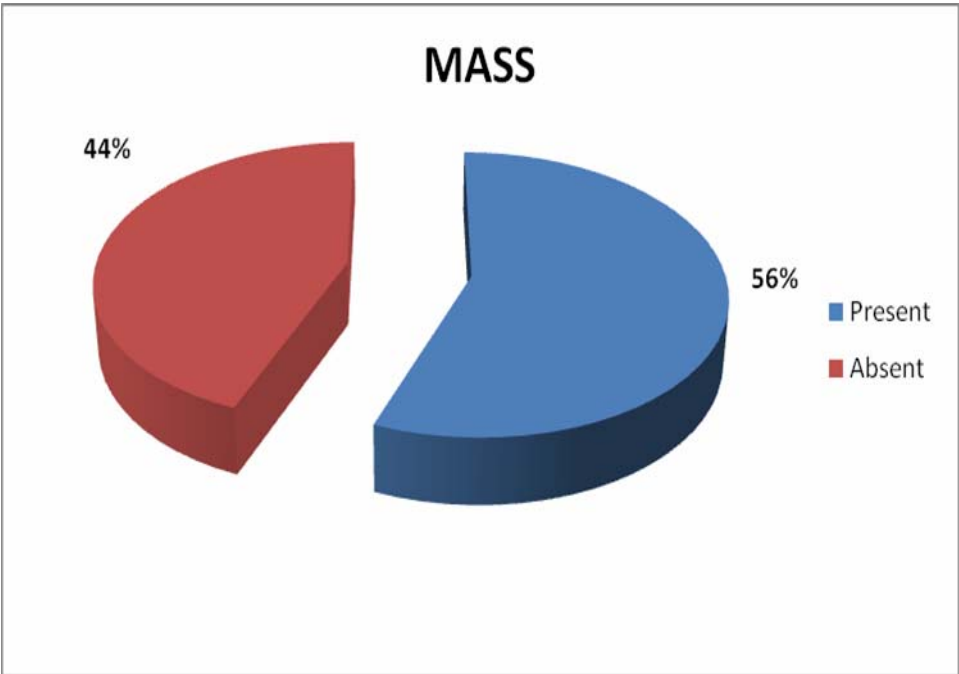
ANTERIOR SEGMENT

RAPD	15%
EOM RESTRICTION	45%
LYMPHADENITIS	2%
IMMATURE CATARACT	12%
CHEMOSIS/FULLNESS	5%
SALMON'S PATCH	2%
PTOSIS	7%
WITHIN NORMAL LIMITS	12%



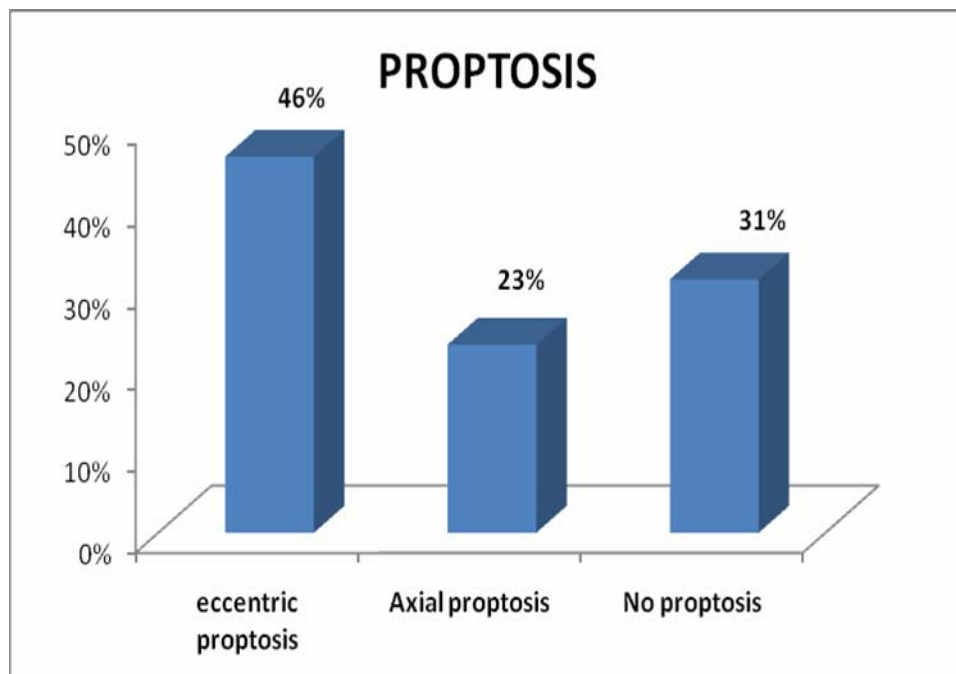
TAB MASS

Mass	
Present	Absent
56%	44%



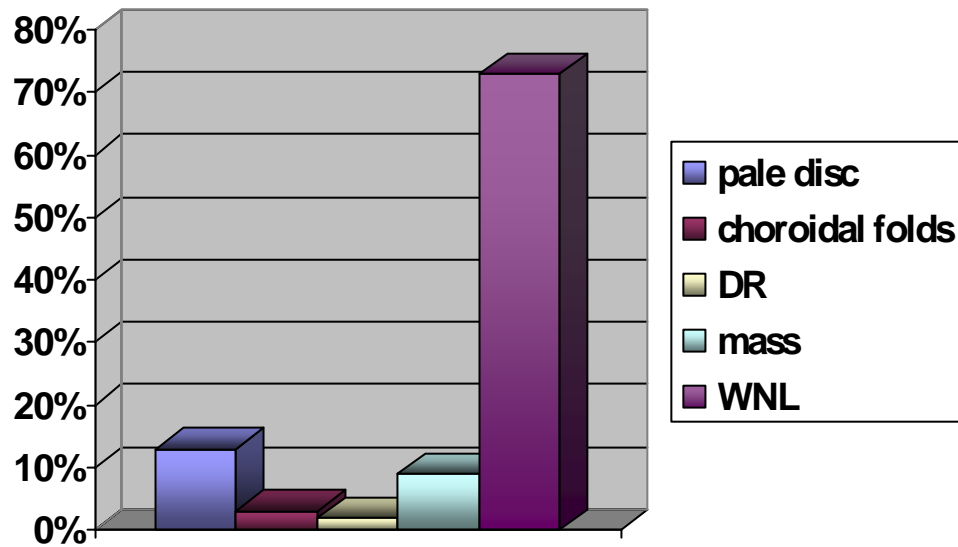
PROPTOSIS

eccentric proptosis	46%
Axial proptosis	23%
No proptosis	31%



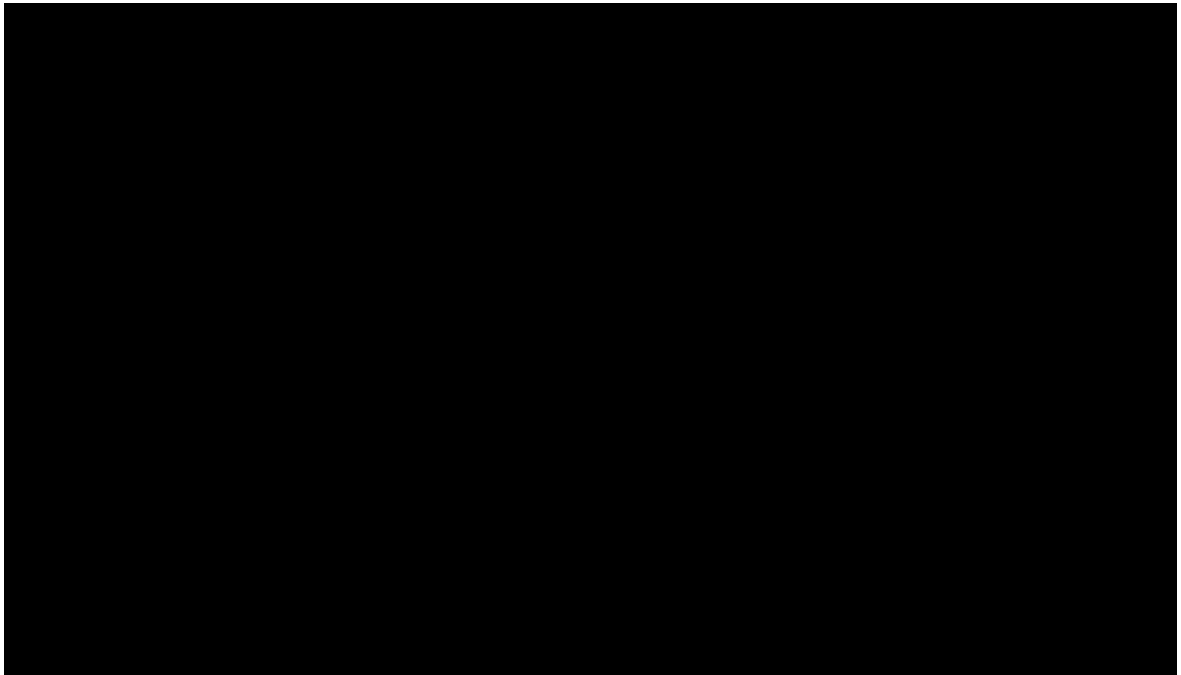
FUNDUS

PALE DISC	13%
DIABETIC RETINOPATHY	2%
CHOROIDAL FOLDS	3%
MASS	9%
WNL	73%



Surgical procedure

Enucleation	10%
Excision biopsy	57%
Incision biopsy	20%
Debulking	10%
Exentration	2%
Orbital decompression	1%

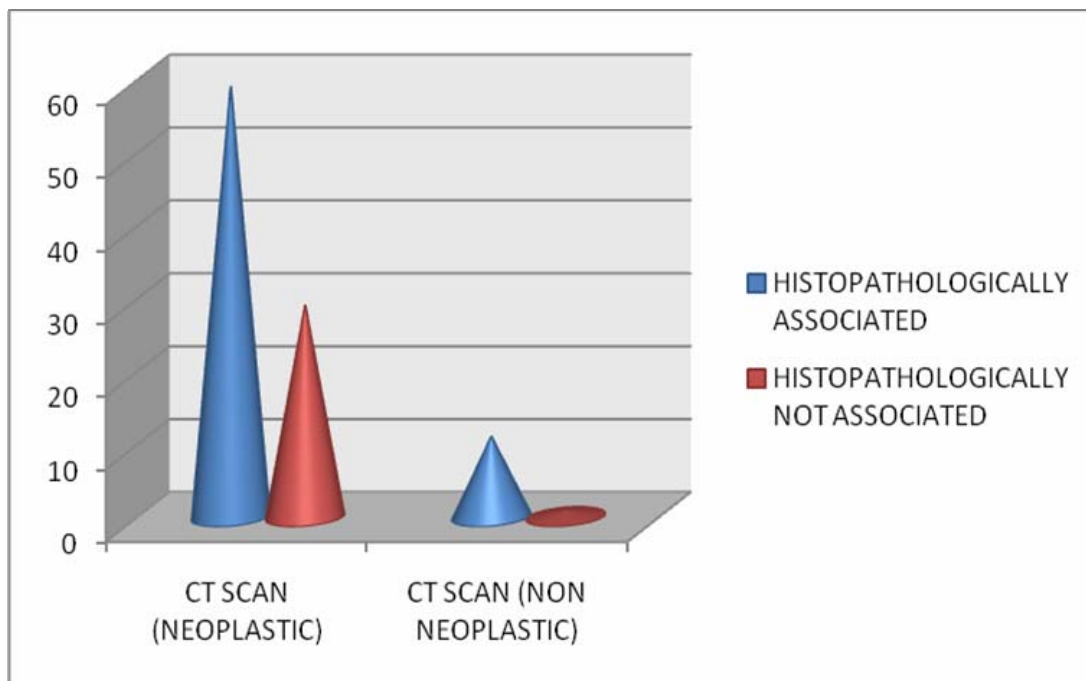


	HISTOPATHOLOGICALLY ASSOCIATED	HISTOPATHOLOGICALLY NOT ASSOCIATED
CT SCAN (NEOPLASTIC)	59	29
CT SCAN (NON NEOPLASTIC)	11	1

CHI- SQUARE VALUE=3.83(P<0.05)

SENSITIVITY=84%

SPECIFICITY=3%



DISCUSSION

In our study, maximum number of patients fell in middle age group, followed by elderly population. There was no sex predilection. 50% were male and 50% were females. There was no definite laterality. 48% of patients had right eye involvement and 48% had left eye involvement. Bilateral involvement was seen in 4% of patients.

Majority of the patients complained of prominence (52%) though a small majority did not notice it (17%). In the above category, some patients had only prominence and some had defective vision along with prominence. 7% of patients presented with double vision, 10% of patients complained of pain. Most of the patients with retinoblastoma presented with white reflex.

Extra ocular movement limitation was a predominant anterior segment finding (45%). RAPD was seen in 15%. 2% presented with lymphadenitis. Salmon patch was seen in 2% of patients. Ptosis was seen in 7% of patients.

56% of patients presented with mass. Patients with both extra-ocular and intra-ocular mass were included.

Totally 69 patients presented with proptosis (46 patients had eccentric proptosis and 23 of them had axial proptosis).

Fundus examination revealed that 13% of patients had pale disc, 3% had choroidal folds. 9% had intra ocular mass. Diabetic retinopathy was seen in 2% of patients.

Majority of patients underwent excision biopsy(57%), followed by incision biopsy(20%). 10% patients underwent enucleation and debulking. 2% were taken up for exentration. One patient underwent orbital decompression.

In 80% of cases, radiological diagnosis by CT scan matched with histopathological diagnosis. Amongst which 59% were neoplastic and 11% were non neoplastic (like aspergillosis)

CONCLUSION

In 80% of cases, radiological diagnosis by CT scan matched with histopathological diagnosis. Chi-square value was 3.83% ($p < 0.05$), which proves a significant association between CT scan and histopathology. Sensitivity was found to be 84%, it was especially high in cases like pleomorphic adenoma, cavernous hemangioma, Non-hodgkins lymphoma, fungal infection and sinonasal malignancy. But, however, specificity was less (3%).

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DIAGNOSTIC VALUE OF CT SCAN IN ORBITAL DISEASES WITH HISTOPATHOLOGICAL CORRELATION

Name

Age

Sex

MRNO

COMPLAINTS

1. Defective vision - RE/LE

- distance/near
- duration
- gradual/sudden
- painful/painless

2. Proptosis

- U/L IBIL
- gradual /sudden
- progressive/stationary/regressive
- variability with posture/sneezing/cough

3. Diplopia

yes/no

4. Pain-eyes

yes/no

5. Headache

yes/no

6. Fever

yes/no

7. Loss of appetite

yes/no

8. Loss of weight

yes/no

9. Nasal discharge

yes/no

10. Bleeding tendency

yes/no

PAST HISTORY

H/o surgery-ENI/thyroid/neurological

H/o remission

GENERAL EXAMINATION

Thyroid swelling	yes/no
Cafe-au-lait spot	yes/no
Shagreen patch	yes/no
Purpuric spot	yes/no
Anemia	yes/no
Lymphadenopathy	yes/no

OTHER SYSTEMS

CVS

RS

ABDOMEN

CNS

LOCAL EXAMINATION

VISUAL ACUITY	-RE LE
LIDS	-erythema/edema/dilated vessels/ retraction/lag/s-shaped
CONJUNCTIVA	-normal/congested
CORNEA	-normal/exposure keratitis /enlarged nerves
ANTERIOR CHAMBER	-quiet/reaction
PUPIL	-size -shape -reaction-normal/sluggish /RAPD
LENS	-clear/cataract
EOM	-full/restricted
VISIBLE MASS	-yes/no
PROPTOSIS	-axial/eccentric -measurements

PALPATION

ORBITAL RIM
INSINUATION
RETROPULSION
REDUCIBILITY
COMPRESSIBILITY
THRILL
PULSATION

-tenderness/erosion/bony defects/thickening

MASS

-size/shape/consistency/warmth/tenderness

REGIONAL NODES

-pre-auricular/sub-mandibular/cervical

AUSCULTATION

-bruit

FUNDUS

FIELD

IOP

RETINOSCOPY

SUBJECTIVE REFRACTION

CLINICAL DIAGNOSIS

INVESTIGATIONS

HEMATOLOGICAL

CT FINDING

-INTRACONAL/EXTRACONAL /CONAL

-INVOLVEMENT OF ORBITAL
MUSCLE/CAVERNOUS SINUS/SOF/IOF

-HYPODENSE/HYPERDENSE/ISODENSE

-CALCIFICATION

ENHANCEMENT WITH CONTRAST

POOR/MILD/MODERATE/BRILLIANT

-BONY DESTRUCTION

-ENCROACHMENT INTO CAVERNOUS
SINUS PARANASAL SINUSES/INTRA-
CRANIUM

CT BRAIN

-BASIFRONTAL
(PARASELLAR/SELLAR/SUPRASELLAR)
-BRAINSTEM
-SKULL VAULT
-VESSELS

SURGICAL TREATMENT

MEDICAL TREATMENT

HISTOPATHOLOGY

MACROSCOPIC

MICROSCOPIC